Chapter 14

CARDIAC TUMORS

Mehmet Şenel BADEMCİ[1]

Introduction

Primary or metastatic cardiac tumors are uncommon in the overall population (1). Historically, these tumors were first described by Malpighi et al. (2) in 1666 as a heart polyp. Later on, in 1934, Barney et al. (3) defined it as a cardiac tumor, as assessed by biopsy materials from a metastatic nodule. Thanks to the advances in the field of cardiac surgery, the first operation for a cardiac tumor, namely excision of a left atrial myxoma, was performed under cardiopulmonary bypass in 1954 (4). With the introduction of echocardiography, a non-invasive imaging modality, into the routine clinical practice facilitated the diagnosis

Classification

Cardiac tumors can be classified as primary and secondary tumors of the heart. The incidence of primary benign cardiac tumors is up to 70%, being higher compared to malignant tumors (5). Among all cardiac tumors, myxoma is the most common primary tumor of the heart, and rhabdomyomas are the most frequent primary cardiac tumors in childhood, usually affecting children aged under the age of 1 (6). On the other hand, based on the autopsy series, the incidence of metastatic tumors is 30-fold higher compared to primary cardiac tumors (7). The most common primary and secondary tumors of the heart are listed in Table 1 (8-9). However, clinical signs and symptoms are based on the localization of the tumor, but not its type (6).

Table 1: *Classification of cardiac* tumors

Primary Cardiac Tumors		**Secondary cardiac tumors**
Primary benign tumors Myxoma Rhabdomyoma Papillary fibroelastoma Lipoma Fibroma Primary malignant tumors Angiosarcoma Rhabdomyosarcoma Mesothelioma and fibrosarcoma		Metastatic cardiac tumors Carcinomas Malignant melanoma Lymphomas Leukemia Lung and breast cancers Infradiaphragmatic tumors

[1]Medeniyet University School of Medicine, Department of Cardivascular Surgery, Goztepe Education and Training Hospital, Istanbul, Turkey

Rhabdomyosarcomas

Rhabdomyosarcomas are the second most common primary tumors of the heart with a malignant nature and usually originate from the right atrium (unlike angiosarcomas which originate from mesenchymal cells), although they can be also located in any parts of the body. The life expectancy is short in patients with rhabdomyosarcomas; however, the success rate increases with chemotherapy and radiation therapy (30).

Secondary (Metastatic) Cardiac Tumors

Secondary tumors are the most frequently seen tumors of the heart caused by the cardiac invasion of the malignant tumors (carcinomas) due to leukemia, lymphoma, malignant melanoma, lung cancer, and breast cancer. Clinical presentation widely varies, depending on the invasion site and size of the tumor. Management of the primary tumor is the mainstay of the treatment. Pericardial effusion is often seen due to cardiac invasion. Surgical treatment options are limited, except for drainage of pleural effusion. Depending on the type of the primary tumor, chemotherapy and radiation therapy can be added to the treatment (6).

Infradiaphragmatic Tumors

Infradiaphragmatic tumors are mainly metastatic neoplasms of renal carcinomas to the right heart. Although they may be asymptomatic, they may lead to certain complaints such as hypertension, dyspnea, and chest pain. Response to chemotherapeutic agents is relatively low than metastatic tumors. Although novel immunotherapeutic agents have been used recently, the prognosis is still poor. In the majority of patients, surgical resection of the tumoral mass is ineffective (31).

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